

Intranodal Palisaded Myofibroblastoma: A Case Report

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ABSTRACT

Intranodal palisaded myofibroblastoma is a rare benign soft tissue tumor almost always arising from inguinal lymphnodes. It usually present as a painless, slow growing inguinal mass. We report herein a case of an intranodal palisaded myofibroblastoma occurring in a 43-year-old man along with its salient clinicopathologic features and differential diagnosis.

Key words: *Soft tissue tumor, Intranodal, Myofibroblastoma*

INTRODUCTION

Intranodal palisaded myofibroblastoma (IPM) also called as intranodal haemorrhagic spindle cell tumour with amianthoid fibre which is a rare benign tumour of lymphnode that may arise from myofibroblasts or smooth muscle cells. It is characterized by the proliferation of hemosiderin laden histiocytes, spindle cells and amianthoid fibres in the lymphnodes. Genetic evidence associated with viral agents with excessive expression of cyclin D1 suggest viral oncogenesis of these lesions¹. The definitive origin of IPM has remained elusive for the following reasons. First, both myoid cells, which are believed to give rise to lymph node blood vessels, and myofibroblasts are found in higher concentrations in inguinal lymph nodes than in other lymph nodes. Thus, these 2 cell lines have an equal chance of giving rise to IPM. Second, the immunohistochemical and electron microscopic studies show features of both myofibroblasts and smooth muscle cells. The features consistent with myofibroblastic origin include actin positivity by immunohistochemistry and the presence of rough endoplasmic reticulum and intracytoplasmic densities of smooth muscle myofilaments by electron microscopy. The features consistent with smooth muscle cell origin include the absence of fibronexus junctions, the absence of numerous rough endoplasmic reticula in abundant cytoplasm, and the presence of external lamina with subplasmalemmal density associating with caveolae. The bottom line is that IPM most likely arises from either smooth muscle cells or myofibroblasts².

These tumours have low grade histologic features and show benign clinical course, so they can be

treated by simple excision. However morphological confusion with metastatic and other spindle shaped lesion, making correct identification is very important³. IPM occur almost exclusively in the inguinal region, followed by submandibular region and neck. These tumours have been found in adults ranging in age from 19-78 years and there is a male predominance (1.5:1). There have been about 50 reported cases of IPM since 1989. We report a 43 years old man with IPM located in inguinal region.

CASE PRESENTATION

Clinical Characteristics:

Physical examination of the 43 years old male patient who presented in surgical OPD of Apollo Hospital Dhaka with the complain of painless swelling in the left inguinal region for 2 years revealed a painless nodular lesion and clinically evaluated as lymphadenopathy. The lesion was excised and sent for histopathological examination.

Macroscopic and Microscopic findings:

Grossly, the nodular lesion had a tan solid cut surface with patchy red brown areas. Sections were stained with HE and masson's trichrome (MT) stains. Immunohistochemically the sections were stained with monoclonal antibody against vimentin, SMA, cyclin D1, desmin and S-100. Microscopic examination reveal proliferation of spindle shaped cells admixed with homogenous eosinophilic accumulation, haemosidesin laden macrophages and extravasated red blood cells with areas of haemorrhage (Fig 1). The tumour cells have scant cytoplasm, elongated nuclei and

coarse chromatin pattern. Among the spindle cells, collagen accumulation were recognized as “amiantoid fibres” showing an irregular distribution and forming stellate structure in some areas (Fig 2). The tumour cells showed no pleomorphism, no atypical mitosis or necrosis is seen. The tumour cells were immunoreactive for cyclin D1, desmin and S-100. In light of these results, the case was diagnosed as intranodal palisaded myofibroblastoma.

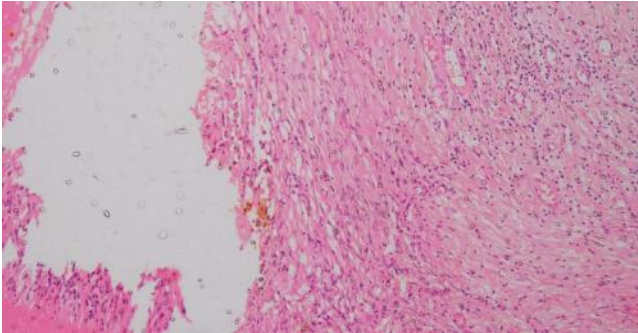


Fig. 1: the tumor showing proliferated spindle cells, zone of sclerosis, haemorrhage and haemosiderin deposit

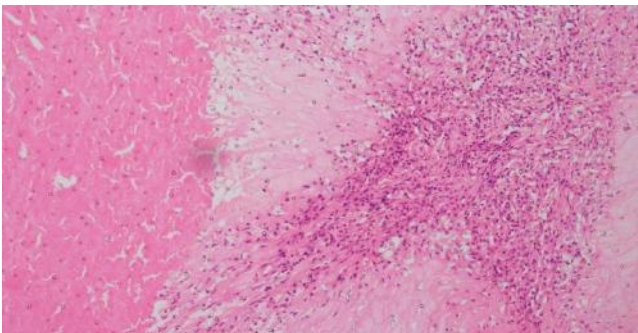


Fig. 2: The tumor showing acellular amorphous material (amiantoid fibres)

DISCUSSION

IPM is a rare neoplasm arising from the stromal component of the lymphnode. Almost all cases develop in the inguinal region lymphnodes, submandibular and mediastinal lymphnodes.¹

IPM produces a slow growing painless inguinal mass with cases mostly commonly seen among age group 35 to 45 years of male, very much similar to present case¹.

Grossly, the cut surface has a solid appearance with irregular haemorrhagic areas. Histopathologically proliferation of spindle cells with nuclear palisading, intraparenchymal

haemorrhage and erythrocytes extravasation, collagen deposits called amiantoid fibres. Immunohistochemically these spindle cells are positive for smooth muscle actin and vimentin.¹

Because cells derived from smooth muscle usually contain desmin the absence of desmin in IPM initially suggested a cell type other than smooth muscle. Intracytoplasmic inclusions, similar to those seen in infantile digital fibroma (an accepted myofibroblastic tumour) seemed to support a myofibroblastic origin. On the other hand, vascular or capsular smooth muscle cells could also give rise to the spindle cells of IPM. A variety of immunohistochemical and ultrastructure features are compatible with either myofibroblasts or smooth muscle cells.⁴

The presence of a spindle cell neoplasm in a lymphnode requires thorough clinical work-up to identify primary neoplasm. Metastatic spindle squamous cell carcinoma, melanoma, sarcomatoid renal cell carcinoma are examples of carcinomas that can present as spindle cell lymphnode lesions. Metastatic sarcoma should also be included. Spindle cell tumours that arise primarily in lymphnodes are Kaposi sarcoma, dendritic cell tumour and benign mesenchymal tumour for example neurofibroma. Nuclear palisading, amiantoid like changes and lack of immunocompromisation distinguish IPM from Kaposi sarcoma. Possibility of metastatic carcinoma, sarcoma and malignant melanoma should be excluded by thorough clinical history and relevant immunohistochemical stains.⁵

IPM are well differentiated tumours with low proliferative activity. They have been agreed to have a benign biological behavior. For this reason, distinguishing these from primary and secondary metastatic malignant lesions of the lymphnode is important.¹

Our case was a middle-aged male patient presenting with an IPM in the inguinal region, the typical location of the tumour. This painless and slow growing lesion was excised and microscopically revealed typical features of IPM. Immunohistochemically the tumour cells were positive for SMA and desmin. Our findings support the suggestion that these tumours arise from differentiated smooth muscle or myofibroblastic cells.

CONCLUSION

In conclusion, IPM a rare tumour should be considered in differential diagnosis of spindle cell lesion of lymphnode because of its ability to mimic metastatic lesion in lymphnode.

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